

Intravenous leiomyomatosis with cardiac extension: Tumor thrombectomy through an abdominal approach

Linda M. Harris, MD, and Constantine P. Karakousis, MD, *Buffalo, NY*

Intravenous leiomyomatosis is an uncommon vascular tumor that may be seen with potentially life-threatening symptoms resulting from intracardiac extension. This tumor is frequently misdiagnosed and treated without appropriate preoperative imaging and planning, which at times leads to inadequate treatment and incomplete resections. The appropriate therapy is complete excision of the tumor. We describe a patient who was treated with a new approach involving a single-stage operation without the need for median sternotomy, cardiopulmonary bypass graft, or hypothermic arrest by resection of the tumor from the point of attachment in the abdominal portion of the inferior vena cava. (J Vasc Surg 2000;31:1046-51.)

Intravenous leiomyomatosis is a rare benign vascular tumor. It is usually confined to the pelvic veins and has been found even less frequently with intracardiac extension. Intravenous leiomyomatosis was first described in 1896, with the first report of intracardiac extension of the leiomyomatosis in 1907 from autopsy analysis.¹⁻³ We report a case of intravenous leiomyomatosis with intracardiac extension and an alternative surgical approach.

CASE REPORT

A 48-year-old white woman was seen by her primary physician approximately 6 months before diagnosis. She had exertional shortness of breath, recurrent palpitations, and dizziness. She had previously been in excellent health before the onset of her shortness of breath. She was a non-smoker with no history of hypercoagulable states, deep vein thrombosis, weight loss, or cancer. Past surgical history was significant for a hysterectomy for benign uterine leiomyoma approximately 3½ years before admission. The results of an electrocardiogram and an echocardiogram performed 6 months before admission were normal. Hypertension subsequently developed, and she underwent a renal ultrasound scan that demonstrated a mass in the inferior vena cava (IVC). Computed tomography

(CT) scan of the chest and abdomen and a transesophageal echocardiogram (TEE) were obtained. A CT showed a mass in the IVC from the level of the renal veins, extending into the right atrium and ventricle (Fig 1). A TEE demonstrated an enlarged right atrium with a large serpiginous mass extending into the ventricle with no discrete point of attachment. Venography was then performed and showed an intraluminal filling defect from the level of the L2 vertebra to the atrium (Fig 2), without occlusion of the IVC. The mass appeared to be attached to the IVC around the level of the renal veins. The patient was taken to the operating room and underwent an anterolateral thoracoabdominal incision with mobilization of the ascending colon. This was done by incising the lateral peritoneal attachments and the duodenum (a Kocher maneuver). The surgical approach is depicted in Fig 3. The right ovarian vein was dilated, was filled with tumor, and was in continuity with the IVC mass. The IVC was then mobilized by ligation of several lumbar veins, and vascular control was obtained inferiorly, below the level of the tumor, and superiorly, below the renal veins with a vessel loop encircling the tumor-filled vena cava. The IVC was mobilized adequately to allow for more superior placement of the vessel loop, if necessary. The right ovary and ovarian vein were excised en bloc. The excised portion appeared rubbery and smooth and did not appear to be friable. The intracaval mass was noted to be firm to palpation but freely mobile within the vena cava. An elliptical incision was made at the junction of the ovarian vein into the IVC. The tumor was adherent to the vena cava just at the junction with the ovarian vein, but was freely mobile after excision of a small ellipse of cava. We decided to extract the tumor without sternotomy, but with cardiac surgery on standby. The tumor was then gently delivered through the venotomy with TEE guidance without difficulty. Postextraction TEE confirmed complete removal of the tumor. Blood loss was less than 100 cc because of

From the State University of New York at Buffalo and Millard Fillmore Hospital.

Competition of interest: nil.

Correspondence: Linda M. Harris, MD, FACS, Department of Surgery, KALEIDA Health/Millard Fillmore Hospital, 3 Gates Circle, Buffalo, NY 14209.

Copyright © 2000 by The Society for Vascular Surgery and International Society for Cardiovascular Surgery, North American Chapter.

0741-5214/2000/\$12.00 + 0 24/4/104601

doi:10.1067/mva.2000.104601



Fig 1. CT scan of the chest demonstrates an intracardiac mass. *CT*, Computed tomography.



Fig 2. Inferior venocavogram shows an intraluminal filling defect that appears to be adherent to the caval wall below the level of the hepatic veins.

cephalad control with a vessel loop around the tumor-filled IVC. A small additional segment of vena cava was debrided and excised to normal-appearing tissue, and the vena cava was primarily repaired. The mass was firm and rubbery in texture without any evidence of friable tissue or tumor clot (Fig 4). The intracaval and intracardiac portions of the tumor measured approximately 31 cm in length with varying widths. Final pathology confirmed the diagnosis of intravenous leiomyomatosis. Histopathology also confirmed a tumor identical to the excised uterine leiomyoma from 3½ years earlier. The patient tolerated the procedure well and was discharged home on the sixth postoperative day. She continues to do well at 6 months' follow-up with no residual tumor and no recurrent cardiac symptoms.

DISCUSSION

Intravenous leiomyomatosis is an uncommon, benign, well-differentiated tumor that may behave in a malignant fashion resulting from venous obstruction or cardiac irritability. The tumor is confined to the vascular channels in contradistinction to benign metastasizing leiomyoma. Although histologically benign, the tumor may recur or cause fatalities that result from venous obstruction and cardiac involvement if not totally excised. Intravenous leiomyomatosis was first identified in 1896 by Birch-Hirschfeld.¹ Durck² and Hormann³ reported the first cases of intracardiac extension of the intra-

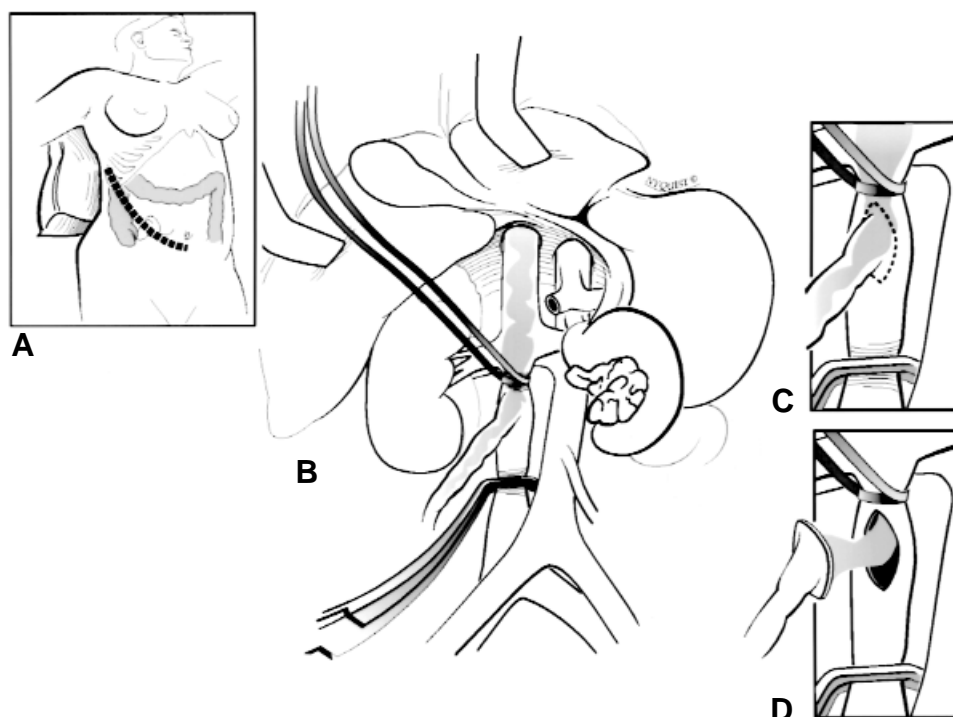


Fig 3. Illustration of surgical approach. **A**, The patient is positioned for an anterolateral approach. **B**, After reflection of the ascending colon and duodenum, the IVC is identified. **C**, Vascular control was obtained with a vessel loop superiorly and a vascular clamp inferiorly. **D**, The tumor was extracted with an ellipse of IVC. *IVC*, Inferior vena cava.



Fig 4. A large, smooth mass with varying widths is seen after extraction from the IVC. *IVC*, Inferior vena cava.

venous leiomyomatosis in separate reports in 1907. The first report in English was not until 1959, by Marshall and Morris.⁴

This disorder is seen exclusively in women, many of whom have undergone previous hysterectomies for uterine leiomyoma. The mean age of a patient with intravenous leiomyomatosis with cardiac extension in the collected series is 48 years (Table).⁵⁻³⁶

Intravenous leiomyomatosis is most commonly seen in the fifth decade in white women with a history of previous pregnancies. A review of the collected series demonstrates that 64% of the women had undergone previous hysterectomies, with a range of 6 months to 20 years before presentation with the intravenous portion of the tumor. Seventy-six percent of the women were postmenopausal; nine per-

Collective review of patients with cardiac extension of intravenous leiomyomatosis

Author/y	Age (y)	Previous hysterectomy	OR approach	Excision	Survival	Cardiac extension
Norris, 1975 ⁵	55	No	—	Partial	—	RA/RV
Tierney, 1980 ⁶	42	Yes (2 y)	CPB, hypothermia	Partial	1 y	RA/RV
Timmis, 1980 ⁷	46	Yes (2½ y)	Sternotomy	Partial	—	RA
Bahary, 1982 ⁸	66	Yes (6 mo)	—	—	Death	RA
	28	Yes (1 y)	CPB—attempt from above	—	Death	RA/RV
Nili, 1982 ⁹	28	Yes (1 y)	CPB—attempt from above	Total	Death	RA/RV
Ariza, 1982 ¹⁰	62	No	Sternotomy, delayed lap	Total	6 mo	RA
Maurer, 1982 ¹¹	52	Yes (15 y)	Sternotomy	Partial	—	RA/RV
Cameron, 1983 ¹²	44	Yes (2 y)	CPB, delayed lap	Total	18 mo	RA
Cleveland, 1983 ¹³	53	—	Hypothermia, combined	Total	—	RA
Iverson, 1983 ¹⁴	51	Yes (7 y)	Hypothermia, combined	Total	1 y	RA/RV
Basso, 1984 ¹⁵	65	Yes (17 y)	CPB, delayed lap	Total	1 y	RA/RV
Politzer, 1984 ¹⁶	46	No	Sternotomy, delayed lap	Partial	—	RA/RV
Gonzalez-Lavin, 1984 ¹⁷	65	Yes (17 y)	CPB, delayed lap	Total	1 y	RA/RV
Mazzola, 1986 ¹⁸	44	Yes (4 y)	Hypothermia, delayed lap	Total	10 mo	RA/RV
Shida, 1986 ¹⁹	42	Yes (5 y)	Sternotomy	Partial	3 yr	RA
		(Same patient)	Hypothermia, combined	Total	8 mo	RA
Garcia, 1986 ²⁰	26	No	CPB, delayed lap	Total	—	RA/RV
Schlosser, 1987 ²¹	34	No	CPB, delayed lap	Total	—	RA/RV
Stegmann, 1987 ²²	53	Yes (10 y)	CPB, delayed lap	Total	6 mo	RA/RV
Roman, 1987 ²³	43	Yes (2 y)	—	—	Death	RA/RV
Rosenberg, 1988 ²⁴	49	Yes (4 y)	Hypothermia, delayed lap	Total	1 y	RA/RV
Kaszar-Seibert, 1988 ²⁵	42	No	CPB, delayed lap	Total	—	RA/RV
Payan, 1989 ²⁶	69	Yes (20 y)	CPB, delayed lap	Total	3 y	RA
Suginami, 1990 ²⁷	72	No	Sternotomy, delayed lap	Total	—	RA
Lee, 1990 ²⁸	38	Yes (2 y)	Combined	Total	3 y	RA
Fukaya, 1991 ²⁹	37	No	CPB, delayed lap	Total	2 y	RA/RV
Cooper, 1992 ³⁰	57	Yes (10 y)	Hypothermia, combined	Total	—	RA
Podolsky, 1993 ³¹	35	No	CPB, combined	Total	—	RA/RV
To, 1993 ³²	54	No	CPB, delayed lap	Total	4 y	RA/RV
Okamoto, 1994 ³³	48	Yes (8 y)	Hypothermia, combined	Total	—	RA
Nakayama, 1994 ³⁴	54	Yes (10 y)	CPB, combined	Total	4 y	RA/RV
Casillas, 1997 ³⁵	48	No	CPB, delayed lap	Total	10 mo	RA/RV
Harris, 1999	48	Yes (3 y)	Single stage—extraction from abdomen	Total	6 mo	RA/RV

Combined, Laparotomy performed simultaneously with sternotomy for tumor extraction; *CPB*, cardiopulmonary bypass; *delayed lap*, laparotomy performed at a separate date after removal of the intracardiac portion through amputation of the tumor at the junction of the IVC with the atrium; *hypothermia*, hypothermic circulatory arrest; *RA*, right atrium; *RV*, right ventricle.

cent had an unknown menopausal status. Presentation is usually related to signs of increased venous obstruction or decreased venous return to the heart. Only two patients in the collected series did not have primary cardiac symptoms: one had abdominal pain,³⁰ and one had a cerebrovascular accident resulting from patent foramen ovale in combination with the tumor.³² The tumors may also be present with sudden death.^{2,3,8,23,36} Unfortunately, because of its rarity, this disorder is frequently misdiagnosed preoperatively.

There are two main theories as to the etiology of these tumors.^{5,37} The first theory maintains that the tumor arises from the vein wall, whereas the second theory suggests that the tumor results from vascular invasion of the myometrium by a leiomyoma. However, the exact etiology of these tumors remains unknown. The tumor is typically a serpiginous,

smooth, solid mass that spreads from the uterine, ovarian, or hypogastric vessels, eventually reaching the IVC and the right atrium.

Differential diagnosis involves primary cardiac tumors, including atrial myxoma, tumor thrombus from other intra-abdominal tumors such as renal cell carcinomas, and primary caval tumors. However, with appropriate preoperative imaging studies, the diagnosis should become more clear.

Because most of the symptoms are related to the cardiac system, echocardiography and electrocardiogram are frequently the initial studies performed. Cleveland et al¹³ have found that although left atrial tumors are essentially exclusively atrial myxomas, right atrial masses are atrial myxomas in only 43% of cases. When a right atrial mass is identified, diagnostic studies should also include venography; CT scan of the chest, abdomen, and pelvis; and trans-

esophageal echocardiography to further delineate the extent and attachments of the tumor. Transesophageal echocardiography will document an intracardiac mass with right atrial and possibly right ventricular involvement with no site of attachment demonstrable. It should also demonstrate extension of the mass from the IVC, thereby ruling out primary cardiac tumors and prompting further studies. Atrial myxomas should have an identifiable site of intracardiac attachment by TEE.

Alternatively, echocardiography may reveal no abnormalities whatsoever when the mass is confined to the IVC. In a patient at risk for intravenous leiomyomatosis who has a normal result from an echocardiogram, further evaluation of the IVC should also be undertaken. The inferior vena cavogram will help to delineate the extent of tumor and at times may demonstrate the origin. If venography and CT show that the tumor adheres to the vena cava, this increases the likelihood of malignant leiomyosarcoma. Venography may also aid in identifying the site of origin for the entrance of intravenous leiomyomatosis into the IVC, thereby improving the surgical plan for inferior control of the vena cava or iliac veins. The CT scan is useful in ruling out other tumors with caval involvement, such as renal cell carcinomas. Uterine leiomyomas may be visible in patients who have not previously had a hysterectomy. Differentiating between benign intravenous leiomyomatosis and primary leiomyosarcoma may still be difficult. Primary leiomyosarcoma may demonstrate a distinct site of attachment to the IVC, as shown in venography or a CT scan. Intraoperative assessment of the attachment of the tumor to the IVC, as well as a history of uterine leiomyomas, may also aid in differentiating the malignant leiomyosarcoma from the benign intravenous leiomyomatosis. With newer imaging modalities, magnetic resonance venography or CT angiography could replace conventional venography and CT scans for the preoperative evaluation of these tumors.

Successful therapy for this tumor involves total excision of the tumor. Excision of the cardiac portion only will result in recurrences that require further surgery or will result in death; such an excision should be avoided.^{5,6,12,19} The first successful extraction of the intracardiac portion of the tumor was reported in 1980 by Timmis et al.⁷ All previously reported intracardiac involvements were fatal. However, this resection resulted in a residual tumor in the IVC. The first successful total resection was reported in 1982 by Ariza et al¹⁰; there was a

delayed laparotomy after resection of the intracardiac portion of the tumor. Since that time, several approaches have been reported for complete removal of the intravenous leiomyomatosis with intracardiac extension. The approaches include either a staged or single operation with cardiopulmonary bypass graft or hypothermic circulatory arrest and total excision of the tumors.⁵⁻³⁵ Reports of attempted removal of the entire tumor from a sternotomy incision alone have been uniformly unsuccessful and fatal, resulting in death from exsanguination.^{8,9} We are the first to report a single-stage procedure with total excision of the tumor without the need for cardiopulmonary bypass graft or hypothermic arrest, by approaching the tumor from its site of attachment within the abdomen. The attachment site for intravenous leiomyomatosis is always intra-abdominal. The site of attachment may include ovarian veins or hypogastric veins and their entrance into the iliac vein or the IVC. Although we approached the tumor with a thoracoabdominal incision, adequate exposure could have been obtained with a purely abdominal incision. This type of approach is not suitable for all intracaval tumors because of the risk of tumor thrombus embolization or tumor fracture. However, the nature of the intravenous leiomyomatosis is such that these tumors are smooth, rubbery tumors without a predilection for tumor thrombus. If there is uncertainty as to the type of tumor or the presence of tumor thrombus or a friable tumor, a combined approach with median sternotomy is certainly warranted. A combined approach with median sternotomy is also recommended for patients with renal cell or adrenal tumors with intracaval and intracardiac extension because of the high risk for tumor fracture or embolization of tumor thrombus. In the case presented, renal and adrenal tumors had previously been excluded, the tumor was clearly freely mobile within the vena cava except for the site of entrance of the ovarian vein, and there was clearly no intracardiac attachment by TEE.

Intravenous leiomyomatosis should be considered in young women with cardiac symptoms and a right-sided intracardiac mass. The level of suspicion should be increased for women with previous hysterectomies for leiomyomas or with pelvic masses. Appropriate preoperative diagnostic testing should be performed to allow for proper surgical planning. Total excision of the tumor is necessary to prevent recurrences and possible fatalities and should be performed in a single or staged procedure. Attempts at removal of the tumor entirely from a sternotomy

without control of the caval attachment site should be avoided. In all of the reported cases in the literature, this tumor is described as a firm rubbery mass without tumor thrombus or regions of friable tissue. This case shows that intravenous leiomyomatosis with cardiac extension can be safely removed without the need for cardiac bypass surgery or circulatory arrest and in a single-stage procedure from the site of caval attachment. Intraoperative visualization of the intracardiac portion of the tumor is important at the time of extraction with TEE.

REFERENCES

- Birch-Hirschfeld FV. Lehrbuch der Pathologischen Anatomie, 5th ed. Leipzig, Germany: F C W Vogel; 1896. p. 226.
- Durck H. Ueber ien Kontinvierlich durch die entere Hohlvene in das Herz vorwachsendes: fibromyom des uterus. *Munchen Med Wehnschr* 1907;54:1154.
- Hormann K. Ueber einen fall von myomatosem uterustumor. *Zentralbl Gynaekol* 1907;31:1604-5.
- Marshall JF, Morris DS. Intravenous leiomyomatosis of the uterus with pelvis: case report. *Ann Surg* 1959;149:126-34.
- Norris HJ, Parmley T. Mesenchymal tumors of the uterus v. intravenous leiomyomatosis. *Cancer* 1975;36:2164-78.
- Tierney WM, Ehrlich CE, Bailey JC, King RD, Roth LM, Wann LS. Intravenous leiomyomatosis of the uterus with extension into the heart. *Am J Med* 1980;69:471-5.
- Timmis AD, Smallpeice C, Davies AC, Macarthur AM, Gishen P, Jackson G. Intracardiac spread of intravenous leiomyomatosis with successful surgical excision. *N Engl J Med* 1980;303:1043-4.
- Bahary CM, Gorodeski IG, Nilly M, Neri A, Avidor I, Garti IJ. Intravenous leiomyomatosis. *Obstet Gynecol* 1982;59:73S-7.
- Nili M, Liban E, Levy MJ. Tricuspid stenosis due to intravenous leiomyomatosis—a call for caution: case report and review of literature. *Tex Heart Inst J* 1982;9:231-5.
- Ariza A, Cerra C, Hahn IS, Shaw RK, Rigney B. Intravascular leiomyomatosis of the Uterus. *Conn Med* 1982;46:700-3.
- Maurer G, Nanda NC. Two-dimensional echocardiographic identification of intracardiac leiomyomatosis. *Am Heart J* 1982;103:915-7.
- Cameron AE, Graham JC, Cotton LT. Intracaval leiomyomatosis. *Br J Obstet Gynaecol* 1983;90:272-5.
- Cleveland DC, Westaby S, Karp RB. Treatment of intra-atrial cardiac tumors. *JAMA* 1983;249:2799-802.
- Iverson LI, Lee J, Drew D, Sharp J, Ecker RR, Young JN, et al. Intravenous leiomyomatosis with cardiac extension. *Tex Heart Inst J* 1983;10:275-8.
- Basso LV, Gradman M, Finkelstein S, Gonzalez-Lavin L. Tricuspid valve obstruction due to intravenous leiomyomatosis. *Clin Nucl Med* 1984;9:152-5.
- Politzer F, Kronzon I, Wiecek R, Feiner H, DeMarco LE, Weintraub PR, et al. Intracardiac leiomyomatosis: diagnosis and treatment. *J Am Coll Cardiol* 1984;4:629-34.
- Gonzalez-Lavin L, Lee RH, Falk L, Gradman MD, McFadden PM, Basso LV, et al. Tricuspid valve obstruction due to intravenous leiomyomatosis. *Am Heart J* 1984;108:1544-6.
- Mazzola A, Gregorini R, Procaccini B, Moretti V, Lucantoni R, Lorenzi W, et al. Intracaval and intracardiac leiomyomatosis of uterine origin. *Ann Vasc Surg* 1986;1:134-8.
- Shida T, Yoshimura M, Chihara H, Nakamura K. Intravenous leiomyomatosis of the pelvis with extension into the heart. *Ann Thorac Surg* 1986;42:104-6.
- Garcia FA, Villanueva RA, Narciso FV, Aventura AP. Intravenous leiomyomatosis of the uterus and pelvis presenting as a cardiac tumor. *Ann Thorac Surg* 1986;42:S41-3.
- Schlosser V, Siebers JW, Kasper W, Bohm N. Vascular tumor in the heart. *J Cardiovasc Surg* 1987;28:132-4.
- Stegmann TH, Garcia-Gallont R, Doring W. Intravascular leiomyomatosis: report of a case and review of the literature. *Thorac Cardiovasc Surg* 1987;35:157-60.
- Roman DA, Mirchandani H. Intravenous leiomyoma with intracardiac extension causing sudden death. *Arch Pathol Lab Med* 1987;111:1176-8.
- Rosenberg JM, Marvasti MA, Obeid A, Johnson LW, Bonaventura M. Intravenous leiomyomatosis: a rare cause of right sided cardiac obstruction. *Eur J Cardiothorac Surg* 1988;2:58-60.
- Kaszar-Seibert DJ, Gauvin GP, Rogoff PA, Vittimberga FJ, Margolis S, Hilgenberg AD, et al. Intracardiac extension of intravenous leiomyomatosis. *Radiology* 1988;168:409-10.
- Payan MJ, Xerri L, Choux R, Gros N, Albrand JJ, Charpin C, et al. Leiomyome geant de la veine cave inferieure. *Ann Pathol* 1989;9:44-6.
- Suginami H, Kaura R, Ochi H, Matsuura S. Intravenous leiomyomatosis with cardiac extension: successful surgical management and histopathologic study. *Obstet Gynecol* 1990;76:527-9.
- Lee PK, David TE, Sloggett C, Ross JR. Intravenous leiomyomatosis with intracardiac extension: an unusual cause of cardiac syncope. *Can Med Assoc J* 1990;142:1257-9.
- Fukaya Y, Iida F, Morimoto M, Nobara H, Miwa H, Nakano H. A case report on successful removal of intravenous leiomyomatosis extending in the right ventricle. *Surgery* 1991;110:909-11.
- Cooper MM, Guillem J, Dalton J, Marboe CC, Corwin S, Todd GJ, et al. Recurrent intravenous leiomyomatosis with cardiac extension. *Ann Thorac Surg* 1992;53:139-41.
- Podolsky LA, Jacobs LE, Ioli A, Kotler MN. TEE in the diagnosis of intravenous leiomyomatosis extending into the right atrium. *Am Heart J* 1993;125:1462-4.
- To WW, Ngan HY, Collins RJ. Intravenous leiomyomatosis with intracardiac involvement. *Int J Gynaecol Obstet* 1993;42:37-40.
- Okamoto H, Itoh T, Morita S, Matsuura A, Yasuura K. Intravenous leiomyomatosis extending into the right ventricle: one-stage radical excision during hypothermic circulatory arrest. *Thorac Cardiovasc Surg* 1994;42:361-3.
- Nakayama Y, Kitamura S, Kawachi K, Kawata T, Fukutomi M, Hasegawa J, et al. Intravenous leiomyomatosis extending into the right atrium. *Cardiovasc Surg* 1994;2:642-5.
- Casillas JA, Lobato J, Llamas P, Meseguer J. Intracardiac extension of intravenous leiomyomatosis: a case report. *J Cardiovasc Surg* 1997;38:83-6.
- Petrow N, Rozynek M. Intravenos wachsendes Uteusmyom. *Zentralbl Allg Pathol* 1970;113:24-31.
- Borland DS, Wotring JW. Intravenous leiomyomatosis of the uterus and broad ligament: report of a case. *Am J Clin Pathol* 1964;42:182-8.

Submitted Jun 9, 1999; accepted Nov 2, 1999.